Pyoderma gangrenosum with unusual periodontal manifestations.

Aruna D.R [1], Sushama G. [2], Pushpalatha [3]

ABOUT THE AUTHORS

1) Dr. Aruna D.R. Professor, Department of Periodontics, V.S.Dental College & Hospital, Bangalore. 560004

2) Dr. Sushama G. Professor, Head of Dept. of Periodontics, V.S. Dental College & Hospital, Bangalore-560004

3) Dr. Pushpalatha, Senior Lecturer, Department of Periodontics, Sree Siddharta Dental College & Research Centre.

Corresponding Author:

Dr. Aruna D.R.

Professor, Department of Periodontics, V.S.Dental College & Hospital, Bangalore. 560004

email: aruvasu@gmail.com

Abstract

Pyoderma gangrenosum (PG) is a rare non infectious ulcerative dermatosis of uncertain aetiology, which is associated with systemic diseases in at least 50% of the patients. It is characterized by recurrent cutaneous ulcerations with mucopurulent or haemorrhagic exudates. The legs are most commonly affected but other parts of the skin and mucous membrane may also be involved. Oral involvement is very rare. We herein present a case of periodontal manifestation of pyoderma gangrenosum in a young woman, which would be the first of its kind.

KEYWORDS: Pyoderma gangrenosum, Oral lesions, cutaneous lesions, periodontal manifestations

Introduction

Pyoderma gangrenosum (PG) is an uncommon ulcerative cutaneous condition of uncertain aetiology. Pyoderma gangrenosum was first described in 1930[1].Clinically it starts with a pustule that rapidly progress and turns into a painful ulcer of variable depth and size with undermined borders. The cutaneous lesions heal by scar formation. The legs are most commonly affected, but other parts of the skin and mucous membranes may also be involved. [2] An underlying disease association is reported in 50-75% of the patients and includes inflammatory bowel disease, rheumatoid arthritis and haematological disorders, the remainder of cases are considered to be idiopathic. The Accurate epidemiological data on PG are missing. The peak of incidence occurs between the ages of 20 to 50 years with women being more often affected than men [2] Cases in infants and adolescents account for only 4% of PG. PG in elderly people has occasionally been reported [3]. The general incidence has been estimated to be between 3 and 10 per million per year .[4]

The 2 primary variants of pyoderma gangrenosum are the classic ulcerative form, usually observed on the legs, and a more superficial variant known as atypical pyoderma gangrenosum that tends to occur on the hands. Patients with pyoderma gangrenosum may have involvement of other organ systems that manifests as sterile neutrophilic abscesses[5]. Intraoral form of PG is pyostomatitis vegetans.[1] Oral involvement is very rare, though very few have been reported in the literature involving buccal mucosa, soft palate and tongue. (J.F.Setterfield). periodontal manifestations of this disorder have never been reported so this would be the first of its kind.

Case Description : A 27 year old female patient was referred from the department of dermatology to the department of Periodontics, Vokkaligara Sangha Dental College & Hospital. Bangalore, with the complaint of painful gums since 2-3 days, bleeding gums and bad breath since 2-3 years which increased during the menstrual period.

Medical history revealed that the patient was a known case of Pyoderma gangrenosum (PG) diagnosed eight years ago with lesions on her lower limbs. Initially she was prescribed Dapsone and Cyclosporine for about a year, after which there was remission. However the patient reported the recurrence of the lesions about 4 years ago and is on steroid therapy since then (omnacortil 20mg).

The patient was moderately built, malnourished, and febrile with right submandibular lymphadenopathy. The patient was anemic and healed PG lesions with scar formation were seen around the nails and in the lower leg region. (fig 1)

Intra oral examination revealed poor oral hygiene with abundant amounts of plaque and calculus. Gingiva was erythematous and fragile with missing interdental papilla w.r.t 42 43 mimicking acute necrotizing ulcerative gingivitis (ANUG), but no typical punched out papilla or psuedomembrane was evident (fig 2). Generalized gingival recession with clinical attachment loss ranging from 4-6mm, pocket depth of 2-3mm and grade II and III furcation involvement w.r.t 46 and 27 with grade III mobility w.r.t 11, 12, 13 was present(fig 3).

Radiological examination revealed generalized angular bone loss with grade II and III furcation involvement w.r.t 46 and 27 (fig 4).

Haematological investigations revealed normocytic hypochromic anemia (Hb10.8%) with severe neutropenia (9%), lymphocytes (90%) and Serological investigation cytoplasmic - Antineutrophil cytoplasmic antibodies (c-ANCA) was positive.

Diagnosis of PG relies on clinical signs first and is

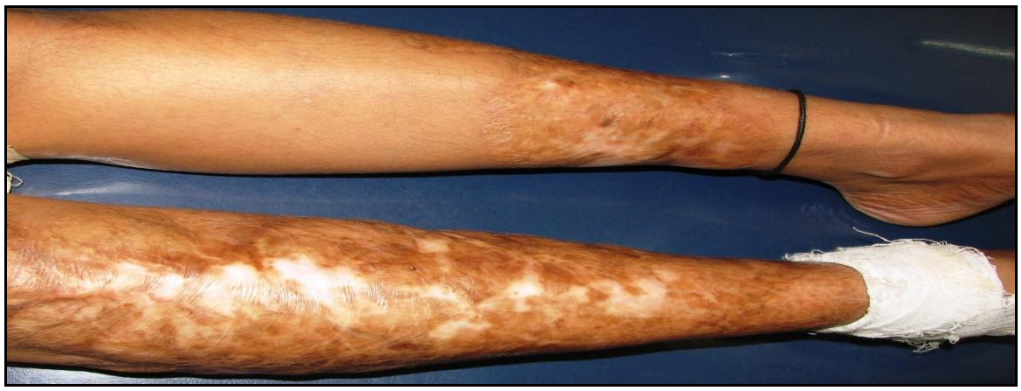

Fig 1. Classic Healed lesions of PG on the lower leg

Fig 2. ANUG like lesion w.r.t 42

Fig 3. Poor oral hygiene Generalized clinical attachment loss

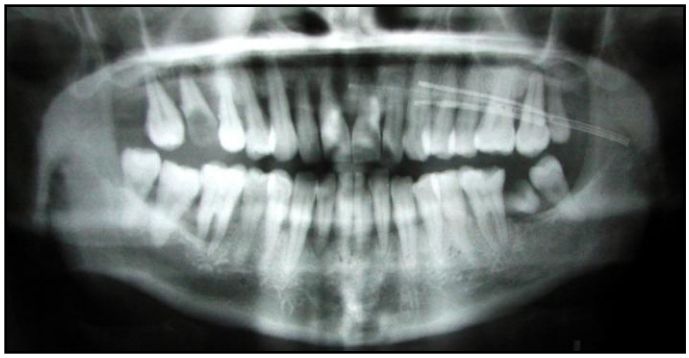

Fig 4. Generalized angular bone loss with furcation involvement

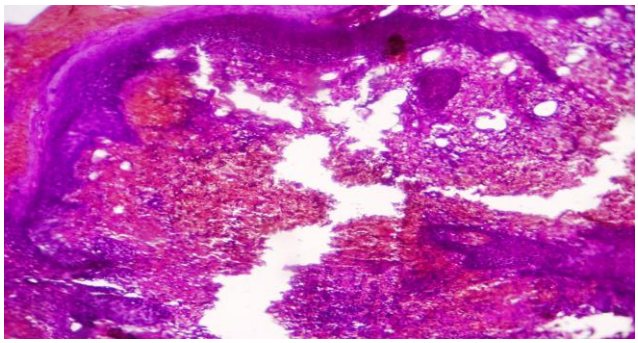

Fig 5. H&E staining of the lesion

supported by biopsy for histopathology. Knowledge of the patient's history for possible underlying disease and specific investigations based on that background are necessary. Therefore diagnosis is made by exclusion of other possible disorders. No laboratory parameter for PG is available. [7, 8]

The histopathology of PG is nonspecific and changes with the stage of lesion. The initial lesions show a deep suppurative folliculitis with dense neutrophilic infiltrate. In about 40% of cases, leukocytoclastic vasculitis is present. PG with [necrotizing] granulomatous inflammation has been described. These reports illustrate the difficulties of a diagnosis based solely on histopathology since concomitant occurrence of PG and systemic necrotizing vasculitis has been observed. [2]

In the present case, biopsy over the skin lesion showed a superficial dermis with mixed inflammatory cell infiltration. Perivascular lymphocytic infiltration and large areas of hemorrhage was seen with areas of necrosis. (fig 5)

Thus based on history, clinical, hematological and serological findings a provisional diagnosis of Periodontitis associated with Pyoderma gangrenosum was established. Generalized Aggressive Periodontitis was considered for differential diagnosis.

Patient was prescribed metronidazole (flagyl)-400 mg t.i.d for 5 days and hydrogen peroxide (3%) mouth rinse (1:1) and to continue the steroid treatment for her PG condition. Patient was reviewed periodically at 5, 15 and 30 days and significant improvements in the oral lesions were seen at the end of one month.

At the end of one month hematological investigations revealed Neutrophil 61 %, Lymphocytes 37%, Hb% 11.3%. Erythrocytes were normocytic hypochromic and microcytic.

Periodontal treatment was instituted after one month and included non-surgical therapy and patient is still on maintenance therapy.

Discussion:

Clinical appearance of Pyoderma gangrenosum is that of a burrowing ulcer which heals by scarring.[1,2] The two primary variants of pyoderma gangrenosum are the classic ulcerative form, usually observed on the legs and a more superficial variant known as atypical pyoderma gangrenosum that tends to occur on the hands. Intra oral form of PG is pyostomatitis vegetans.[1]

In differential diagnosis Crohn's disease, Ulcerative colitis, phemphigus vulgaris and cicatrical phemphigoid were ruled out. In the present case patient never reported any of the signs and symptoms of gastro intestinal diseases or conditions such as diarrhea, abdominal pain, rectal bleeding, anorexia, weight loss. Crohns disease and Ulcerative colitis are the most common gastro intestinal diseases which can also have pyoderma like skin lesions. Thus these diseases were only considered for differential diagnosis.

Phemphigus vulgaris is an auto immune disorder which clinically presents as bullae or vesicles which rupture and result in an ulcer, clinically nikolsky's sign can be appreciated and the lesions heal without scar formation.

Cicatricial pemphigoid is an auto immune disease which predominately affects the mucous membranes, including the conjunctiva, and occasionally the skin. Patients with cutaneous involvement present with tense blisters and erosions, often on the head and the neck or at sites of trauma. Most patients with cicatricial pemphigoid are elderly, with a mean age of 62-66 years. Pyoderma gangrenosum commonly affects the skin and it affects patients of 30-50yrs and mainly occurs in the lower leg regions. [7, 8.]

In the present case biopsy over the skin lesion was non specific showing mixed inflammatory cell infiltration in the superficial dermis. Perivascular lymphocytic infiltration and large areas of hemorrhage was seen with areas of necrosis. (fig 5) thus confirming the diagnosis.

Our case was a known case of Pyoderma gangrenosum (PG) on medications. PG most commonly occurs on the lower leg region; in our case also healed lesions of PG were seen on the lower leg region and around the nails. Our patient gave a history of oral lesions on the oral mucosa, tongue but when she reported to the department, no PG like lesions were seen intraorally.

Previously reported cases of prominent oral mucosal involvement in PG are available. Sites of involvement included the tongue, palate, tonsillar fauces and buccal mucosa. [6] No literature is available on the periodontal involvement in Pyoderma gangrenosum (PG) and our case has significant periodontal findings like generalized loss of attachment, vertical bone loss, furcation involvement, mobility. loss of interdental papilla in the lower anterior tooth which mimics acute necrotizing ulcerative gingivitis (ANUG), like lesion, though no clear cut punched out margins were seen.

Pathophysiology of pyoderma gangrenosum is poorly understood, but dysregulation of the immune system, specifically altered neutrophil chemotaxis is believed to the involved [1, 2].

In our case c-ANCA serological test was positive which suggest that antibodies were found against myeloperoxide enzyme resulting poor neutrophil function. This altered neutrophil function could also be the possible cause for rapid periodontal destruction [8, 9].

In a study conducted to assess the role of ANCA in the pathogenesis of adult periodontitis, found that periodontitis associated bacteria have well known polyclonal B-cell activator capacity, and this could be the primary source of ANCA. Gingiva from such patients contains high number of antibody secreting cells mimicking auto anti bodies. Binding of ANCA to neutrophils activates these cells producing degranulation, reactive oxygen species and translocation of ANCA reactive proteins to cell surface, there by exposing these antigens to the immune system. Interaction of these proteins with their cognate antibodies activtes complement lysis and the periodontal destruction. [8]

Regarding the initial neutropenia and lymphocytosis seen may be due to any viral infection which returned to normal after a month. [8, 9]

Patient also reported the flare up of lesions during menstration, so a possible influence of female sex hormones on periodontium making it susceptible to periodontal infections can also be considered as the contributing factor. [10]

Thus it can be concluded that pyoderma gangrenosum is a rare destructive ulcerative skin disorder with dysregulation of immune system particularly neutrophil homeostasis such as altered chemotaxis.

Oral manifestations are rare but it could manifest in periodontium with severe destruction at a young age. So the Dermatologists of such patients should be made aware of possible oral mucosal and periodontal involvement and proper precautionary treatment measures should be implemented as early as possible.

References:

1. J Mark Jackson, Jeffrey P Callen: Pyoderma Gangrenosum emedicine.medscape.com Updated: Oct 30, 2008

2. Uwe Wollina: Pyoderma gangrenosum – a review Orphanet Journal of Rare Diseases 2007, 2:19

3. Graham JA, Hansen KK, Rabinowitz LG, Esterly NB. Pyoderma gangrenosum in infants and children. Pediatr Dermatol. Mar 1994;11(1):10-7.

4. Powell FC, Schroeter AL, Su WP, et al.: Pyoderma gangrenosum: a review of 86 patients. Q J Med 1985, 55:173-186

5. Brown TS, Marshall GS, Callen JP. Cavitating pulmonary infiltrate in an adolescent with pyoderma gangrenosum: a rarely recognized extracutaneous manifestation of a neutrophilic dermatosis. J Am Acad Dermatol. Jul 2000;43:108-12.

6. J.F.Setterfield, P.J.Shirlaw, S.J.challacombe and M.M.Black: Pyoderma gangrenosum associated with severe oropharyngeal involvement and IgA paraproteinaemia Br J Dermatol 2001; 144: 393-396.

7. Norman K Wood,Paul W. Goaz, Differential diagnosis of Oral and Maxillofacial lesions, 5th edition,Elsevier publisher Ltd,2007:71

8. Novo E, Viera N: Antineutrophil cytoplasmic antibodies a missing link in the pathogenesis of periodontal disease. J Periodont Res 1996;31:365-368

9. MW Chia, L Teo, YK Tay, WT Poh: Pustular pyoderma gangrenosum: An uncommon variant which is easily misdiagnosed. Dermatology Online Journal 14 (2): 21.

10. Mariotti A. Sex steroid hormones and cell dynamics in the periodontium. Crit Rev Oral Bio and Med.1994; 5: 95 -124